CASE REPORT

Extraskeltal Ewing’s sarcoma of scalp

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Abstract

Extraskeltal Ewing’s sarcoma is a rare tumor and has to be kept in mind in the differential diagnosis of small cell tumors of soft tissue. It carries significant morbidity and mortality if not diagnosed and treated early. This is a case of a 14 yr old female who presented with a progressively growing tender mass over the left side of scalp without bony involvement initially but later on developed right sided body weakness and expired within a month span.

Keywords: Ewing’s sarcoma, Small cell tumours of soft tissue, malignant growth of scalp

Introduction:

Primary Extraskeltal Ewing’s sarcoma of soft tissue are ones in which underlying bone involvement is not found. They are uncommon mesenchymal tumors and have to be differentiated from other small cell tumors on the basis of immuno-histochemistry and electron microscopy. We are presenting a case of this rare tumor in a young female without any predisposing factor or lesion.

Case report:

A 14 years old female initially presented as a OPD case of sebaceous cyst over scalp, excision biopsy of which showed inflammatory cells and was not conclusive. She again presented within a month span with a rapidly progressive growing mass over the left side of the scalp from the previous scar site along with severe anaemia. It was also associated with constitutional symptoms like fever, malaise and fatigue. On examination patient was of average built with severe pallor. No evidence of bony metastasis to skull, ribs, vertebrae or lungs in the form of hemoptysis or chest pain was present. Local examination revealed a huge tender mass of about 10 x 10 cm over the left side of scalp involving frontal, temporal, parietal and almost whole of the occipital region. It was soft in consistency, tender, immobile with gangrenous and necrotic patches over its surface. It was friable and bled on touch. Hemogram with ESR revealed severe anaemia with a Hb of 6. Other blood tests including renal function test, liver function test, serum calcium and serum alkaline phosphatase were within normal limits. CT scan of brain was done which showed an extracranial mass with no intracranial extension. As tumor was too huge it was decided to go directly for Wide local excision instead of doing excision biopsy first there fore whole of the mass was excised up to the periosteum of cranium and was sent for histopathological examination. On microscopic examination, spindle shaped cells loosely arranged in vague fasicles and sheets with interspersed fine fibrovascular connective tissue. The cells showed faint eosinophilic to clear cytoplasm with indistinct boundaries. The nuclei are round to oval showing open chromatic with small nucleoli. Brisk mitotic activity noted about 32 mitotic figures/10HPF. Tumor cells were positive with PAS stain. Final diagnosis required Immuno histochemistry which showed positivity against Mic 2(CD 99) and S-100 hence confirmed the diagnosis of Ewing’s sarcoma.

Our next plan was skin grafting followed by chemotherapy which was planned two weeks after initial excision but peroperatively there was an evidence of local recurrence due to which graft-
ing couldn’t be done. A day before the commencement of chemotherapy patient developed right sided body weakness and expired. Accurate cause couldn’t identified but it was thought to be due to intracranial involvement.

Discussion:
Extraskeletal Ewing’s sarcoma is a rare entity. Ewing’s sarcoma develops from a type of primitive nerve cell and may occur in the soft tissues of the body as well as in bone. Most common sites for Ewing’s Sarcoma are bones (47%), pelvis (19%), ribs (12%). Ewing’s sarcoma of Skull vault is very rare and makes up less than 1% of all brain tumors. The most common presenting complaint is a visible or a palpable mass in children out of which some are tender on palpation. EES is usually aggressive with a high rate of recurrence. Distant metastases are common, where lung is the commonest site. Although the prognosis for this tumour is grave, it is potentially curable. However prognosis depends on various factors like presence of distant metastases at the time of diagnosis, age greater than 10 years, central location, size and poor response to chemotherapy. Therefore early treatment before metastases is the key to long term survival in patients with Ewing’s Sarcoma.

Conclusion:
Extraskeletal Ewing’s sarcoma is a very rare entity. Very rarely it can develop from a primitive nerve cell or soft tissue of the body as well as in bone. As Ewing’s sarcoma of skull vault is very rare one should diagnose and treat the patient early to get better prognosis.

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